

CEREBERIFORM INTRADERMAL NEVUS

(A case report)

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Summary

A 35 year old woman with cereberiform nevus on the right cheek is reported for its rarity and its occurrence in an uncommon site.

Introduction

Cereberiform nevus is a recently described variant of nevocellular nevus. It is considered to be a non-pigmented variant of giant pigmented nevus. Clinically starting as a skin coloured or brown to black spot it slowly enlarges in size with convolutions reaching the size of several centimeters. Usual age of onset is 5-10 years. A spurt in growth may occur during pregnancy, or after hysterectomy. The ultimate size of the nevus may vary from 2×3.5 cms to 14×13 cms. The common site of occurrence are the parietal, occipital and temporal regions of the scalp. The temporal lesion may extend on to the cheek and pre-auricular regions. Pruritis, burning and alopecia are the predominant symptoms. Bleeding following trauma and infection leading to abscess formation can also occur. Pathologically these nevi will show islands of nevocellular nevus in the upper dermis and neuroid nevus in the deeper dermis. Vascular hyperplasia resembling lymphangioma and atrophy of hair follicles may also occur. Junctional

activity is usually absent in a well developed lesion. Occurrence of malignant melanoma and bony defect under the nevus are dreaded complications of this disorder. This nevus has to be differentiated from plexiform neurofibroma and other causes of cutis verticis gyrata. The plexiform neurofibroma lacks in islands of nevus cells and will show a number of nerve fibres with Bodian stain. Surgical extirpation is the treatment of choice for this condition (Fig. 1 & 2).

Case Report

35 years old lady presented with skin lesions of 5 years duration on the right cheek. It started as a pruritic nodule and was slowly increasing in size. Excepting for mild itching there was no other symptom. Family history was negative for such lesion. Examination revealed a convoluted cereberiform plaque with nodular surface involving whole of right cheek and extending to the lower lid of right eye (Fig. 1). Plaque was uniformly firm, non-compressible and not attached to deep structures. There was no involvement of facial nerve or buccal mucosa. Regional nodes were not enlarged. Routine hematological and urine examinations were normal. X-ray skull revealed no bony abnormality. Biopsy

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Fig. 1

Shows cereberiform nevus on the right cheek.

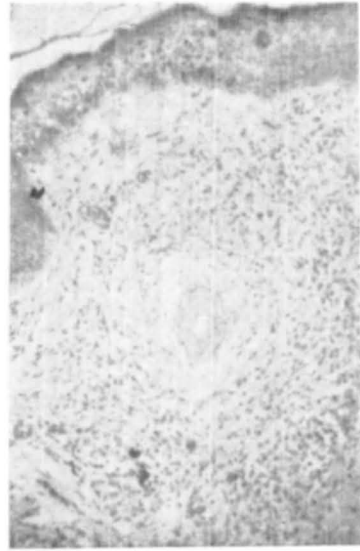


Fig. 2

H & E x 100 shows islands of nevus cells with few pigment-containing cells in the upper and mid dermis.

of the lesion showed islands of nevus cells in the upper dermis and nests of nevus cells with neuroid component in the mid and lower dermis (Fig. 2). Bodian stain failed to show any nerve bundles. Patient was not willing for surgical excision and repair.

Discussion :

In this case the possibility of neuro fibroma was considered. The presence of dermal nevus picture in histopathology and failure to demonstrate nerve bundles with Bodian stain supported the diagnosis of cereberiform nevus. The interesting features of this case are late onset, involvement of cheek and extension onto the eye lid with involvement of the conjunctiva.

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